CC-TGA

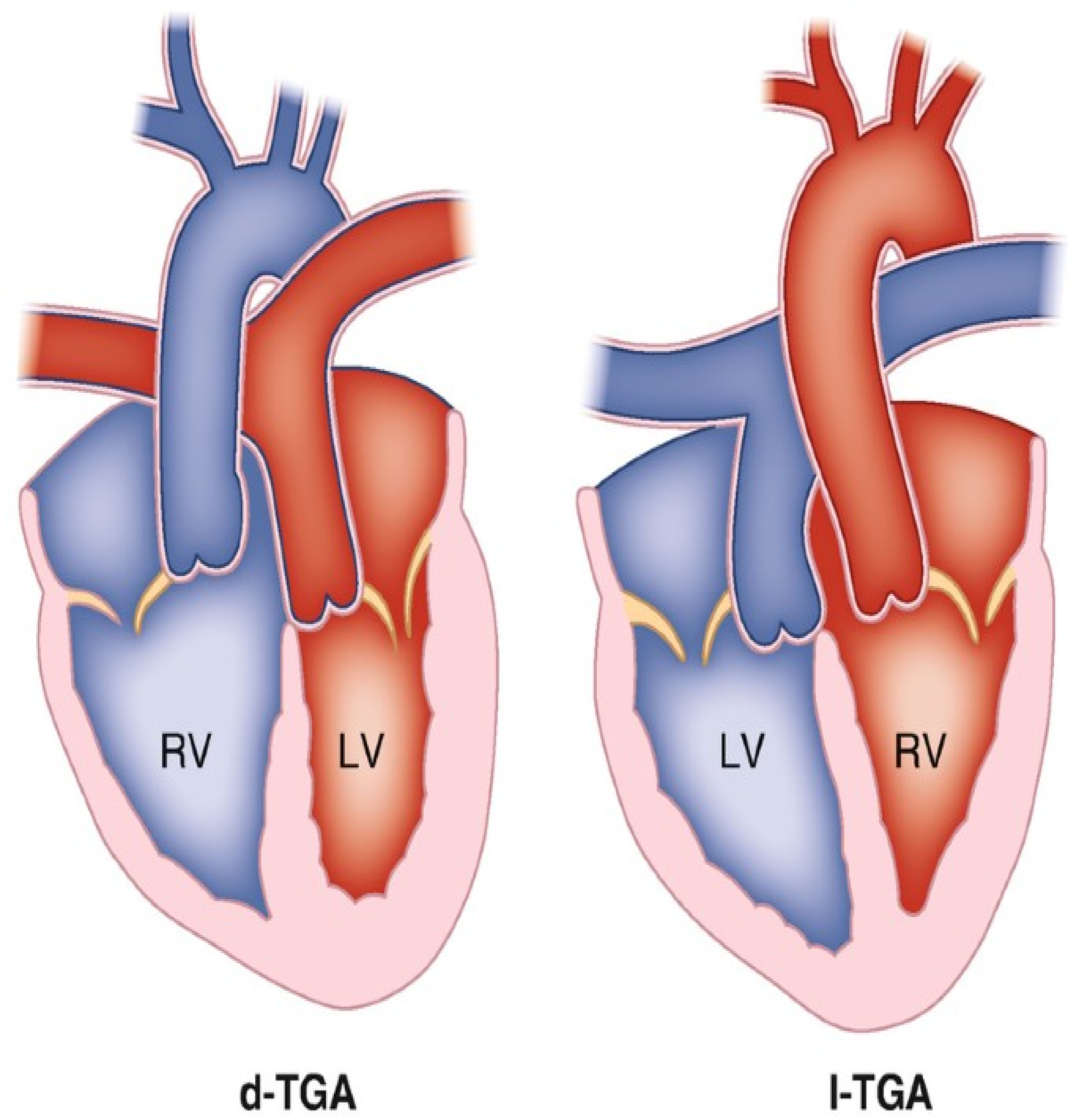
Core Concepts In Diagnosis and Management

Mahmoud Saber, MSc

Transposition

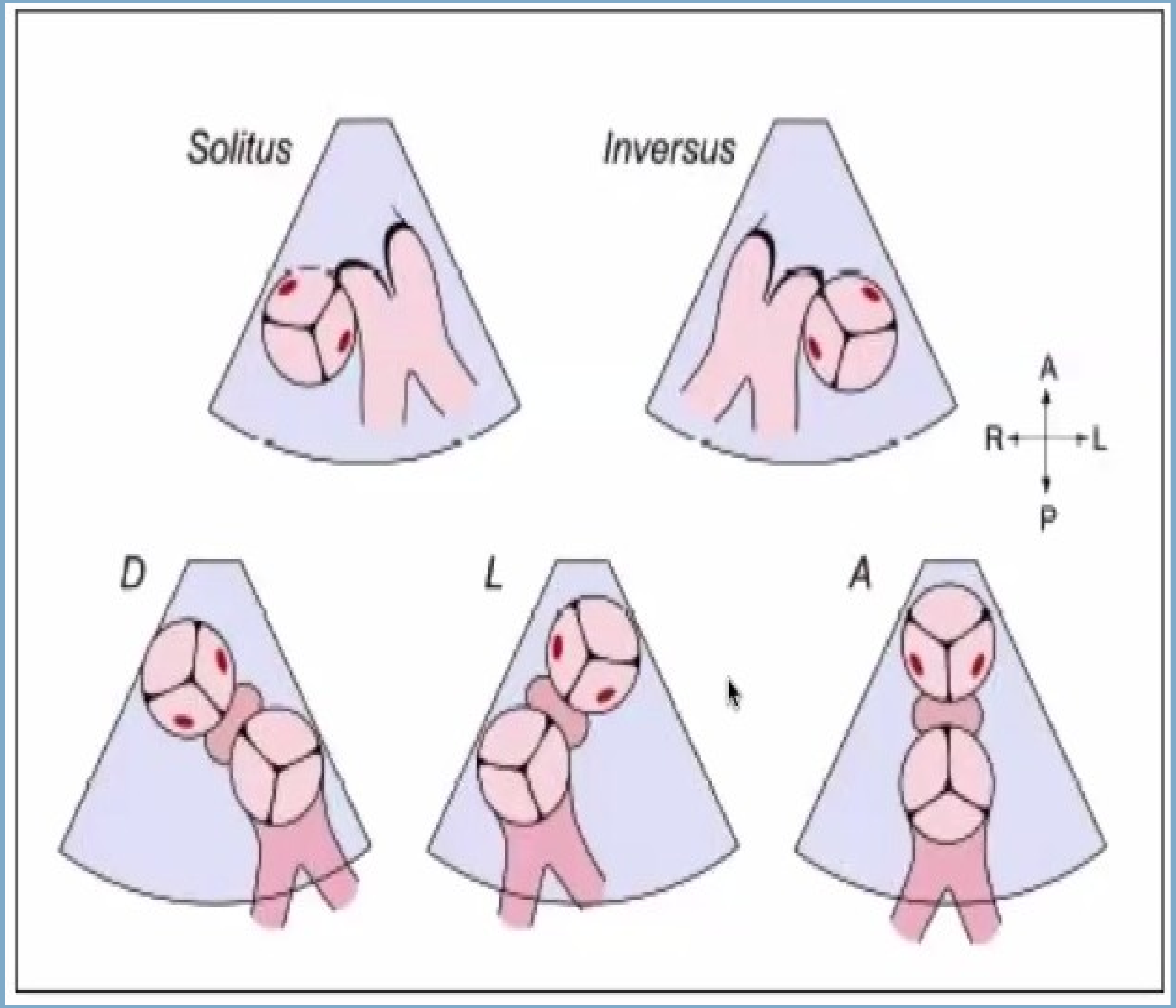
- VA discordance
- Great vessels displaced across the IVS
- Each arterial trunk arise from morphologically inappropriate ventricle

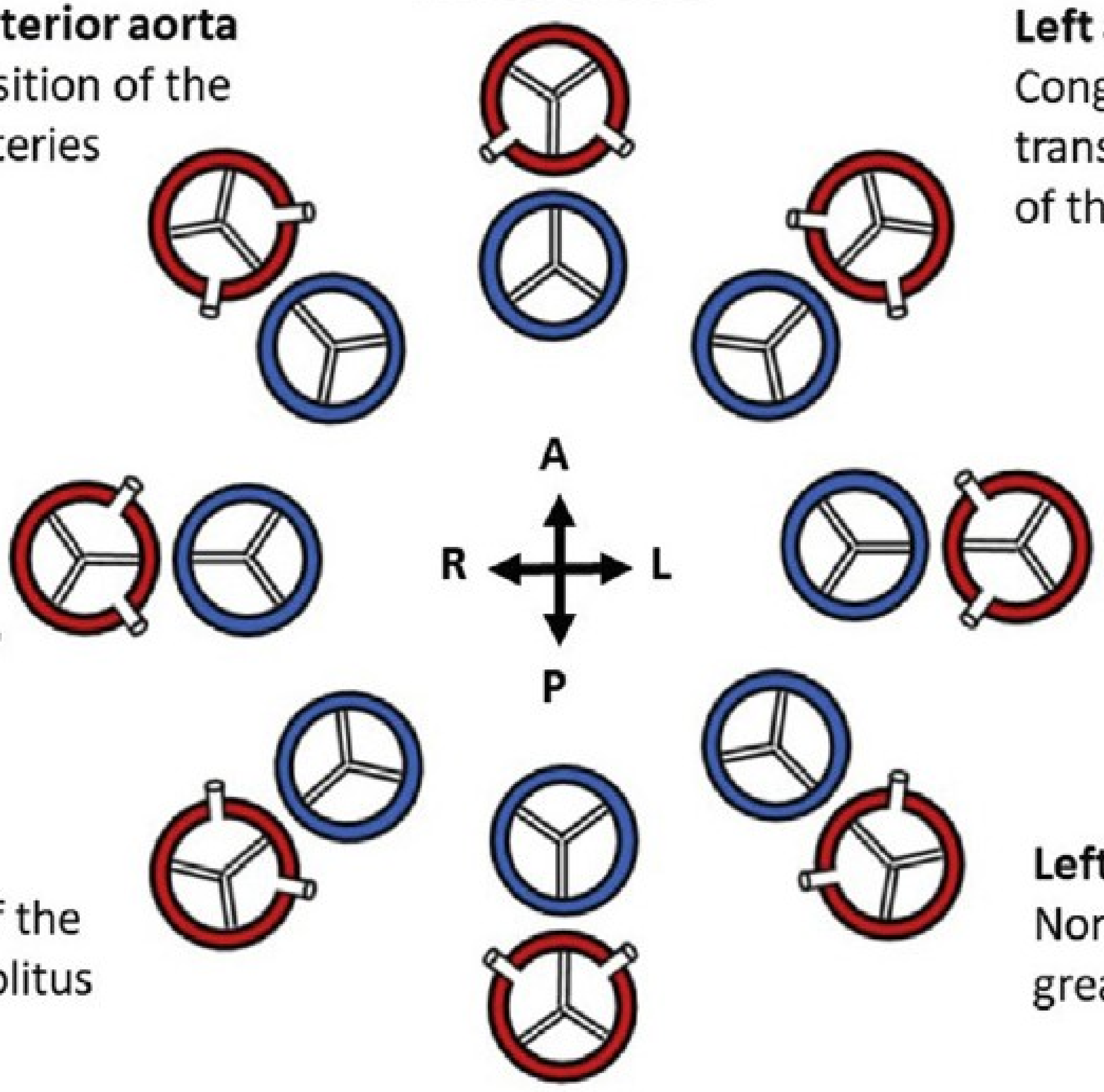
LV >> PA RV >> Ao


RV
LV
LV
RV
d-TGA
l-TGA

Malposition

- Abnormal great vessels arrangement
- But not displaced across IVS.
- VA concordance, DORV .. single outlet


Solitus
Inversus
A
R
L
P
D
L
A



Anterior aorta

Right anterior aorta
Transposition of the
great arteries

Left anterior aorta
Congenitally corrected
transposition
of the great arteries

Side by side great arteries - right aorta
Double outlet right
ventricle ''Taussig Bing''

Side by side great arteries - left aorta

Right posterior aorta
Normal relationship of the
great arteries - situs solitus

Left posterior aorta
Normal relationship of the
great arteries – situs inversus

Posterior aorta

AV Discordance (reversed off-setting AVV)

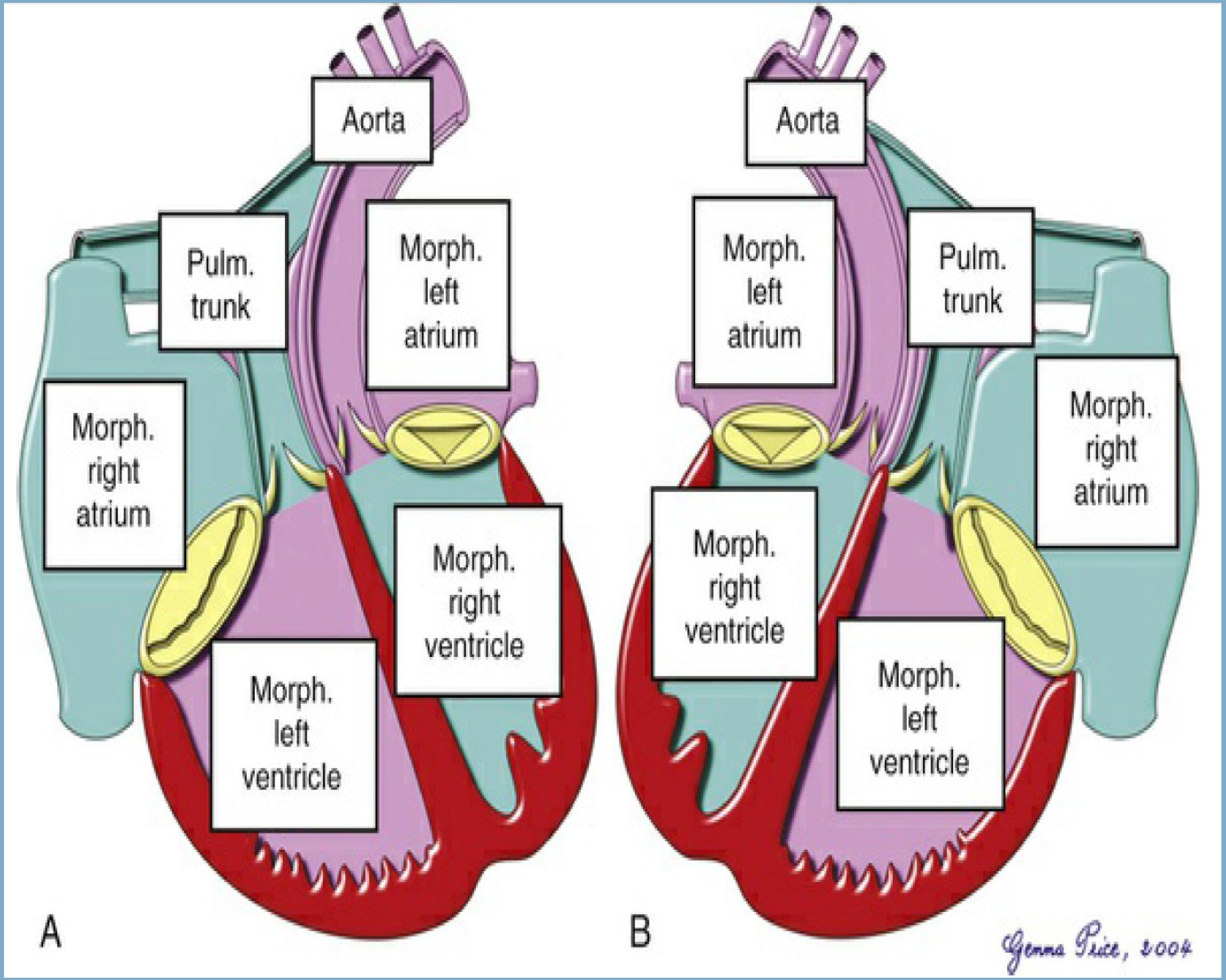
- Transposition (VA discordance)
- Isolated Ventricular Inversion (AV concordance; rare)
- Double Outlet : commonly DORV.
- Pulmonary or Aortic Atresia (CC-TGA with PA?)

CC-TGA:

- Transposition + Ventricular inversion
- Double discordance (AV & VA discordance)
- L-TGA (Levo looping, Left anterior aorta):aorta may be right sided or directly anterior, also left sided aorta may be present in D-TGA.

CC-TGA:

- LA >> mRV >> Ao
- RA >> mLV >> PA
- Systemic RV
- Systemic TV: septal attachment

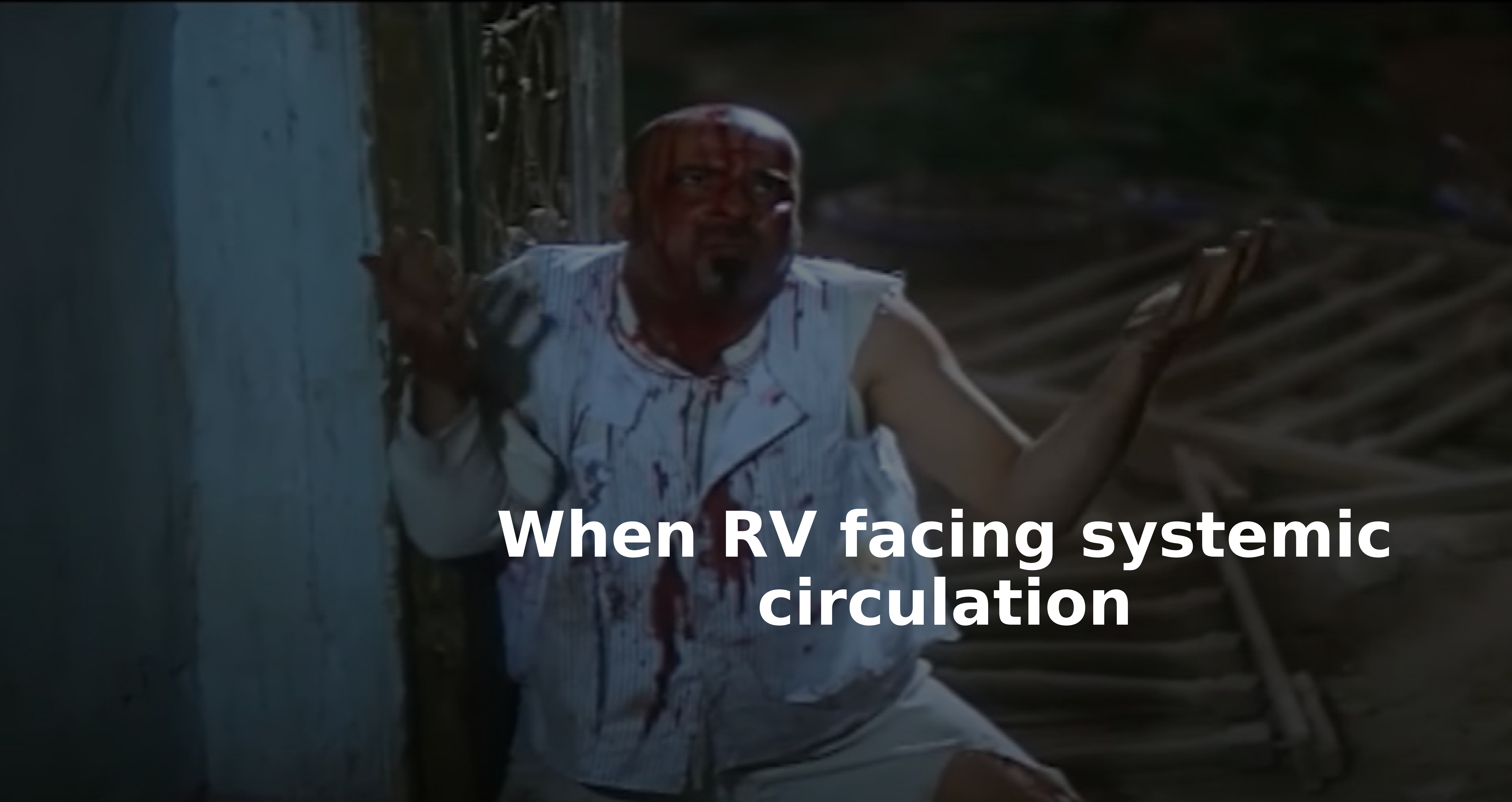

Aorta
Pulm. trunk
Morph. left atrium
Morph. right atrium
Morph. right ventricle
Morph. left ventricle
A
Aorta
Morph. left atrium
Pulm. trunk
Morph. right atrium
Morph. right ventricle
Morph. left ventricle
B
Gemma Price, 2004

Systemic RV & systemic TV:

- Complex interaction between RV dysfunction and TR.
- RV: different shape & different coronary blood supply.
- TV: not designed as well for systemic pressure

  septal attachment >> more dysfunction with

  more RV dilatation



When RV facing systemic circulation

Pathophysiology

- Functionally; corrected, but long-term systemic RV & TR.
- Associations:
    - VSD (~80%): commonly PM (sub-pulmonic)
    - PS (~50%): 80% of these cases combined with VSD

      Variable level, may be combined
    - TV abnormalities (~60-80%): dysplasia & Ebsteinoid.
    - Heart Block (progressive AV block; uncommon at birth) – SVTs.

Clincial Presentation:

- Great variety in age and nature of clinical presentation:
- Associations:

  VSD: HF symptoms – recurrent chest infection – failure to thrive

  VSD & PS or mild forms of PS: balanced

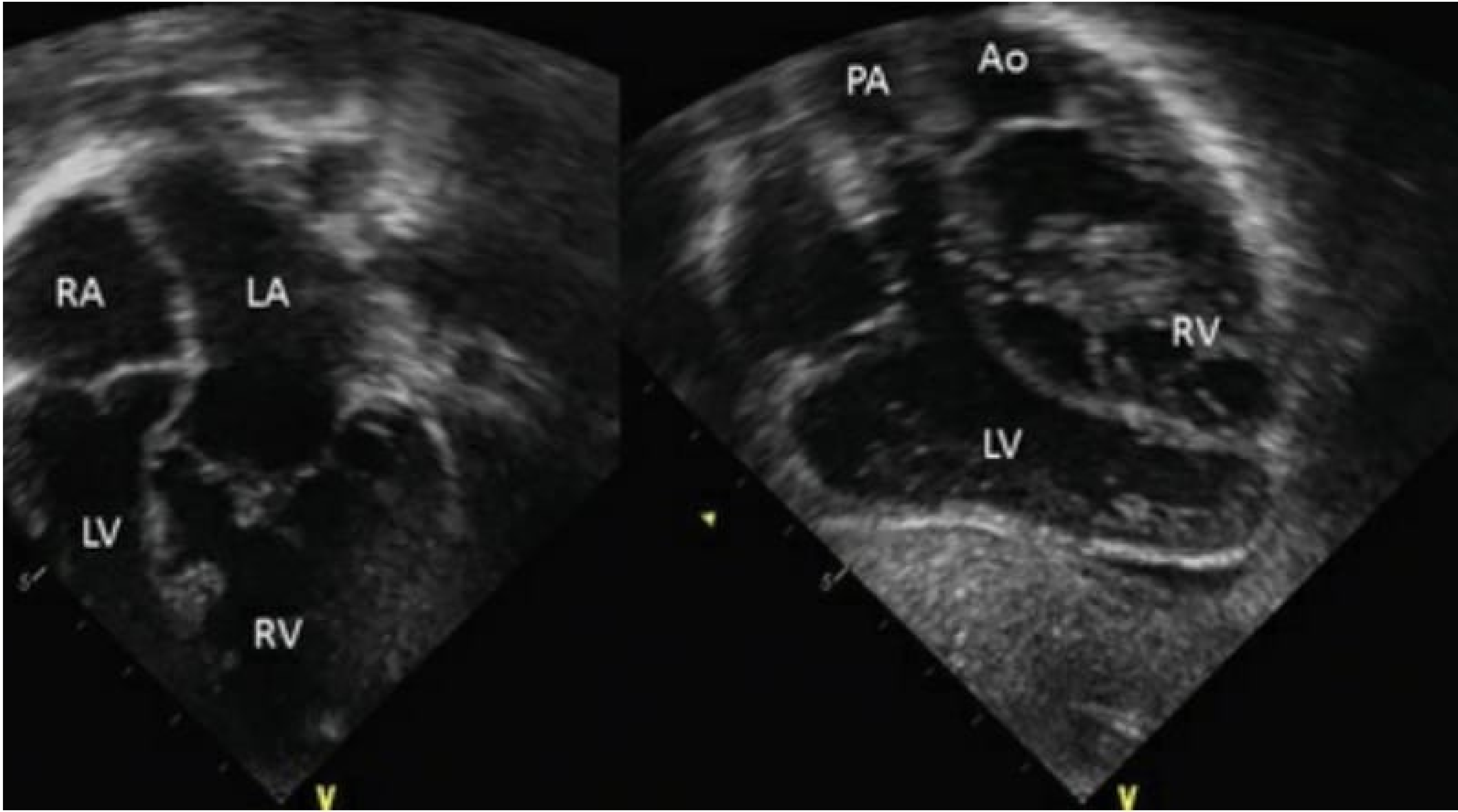
  severe PS or PA: cyanosis

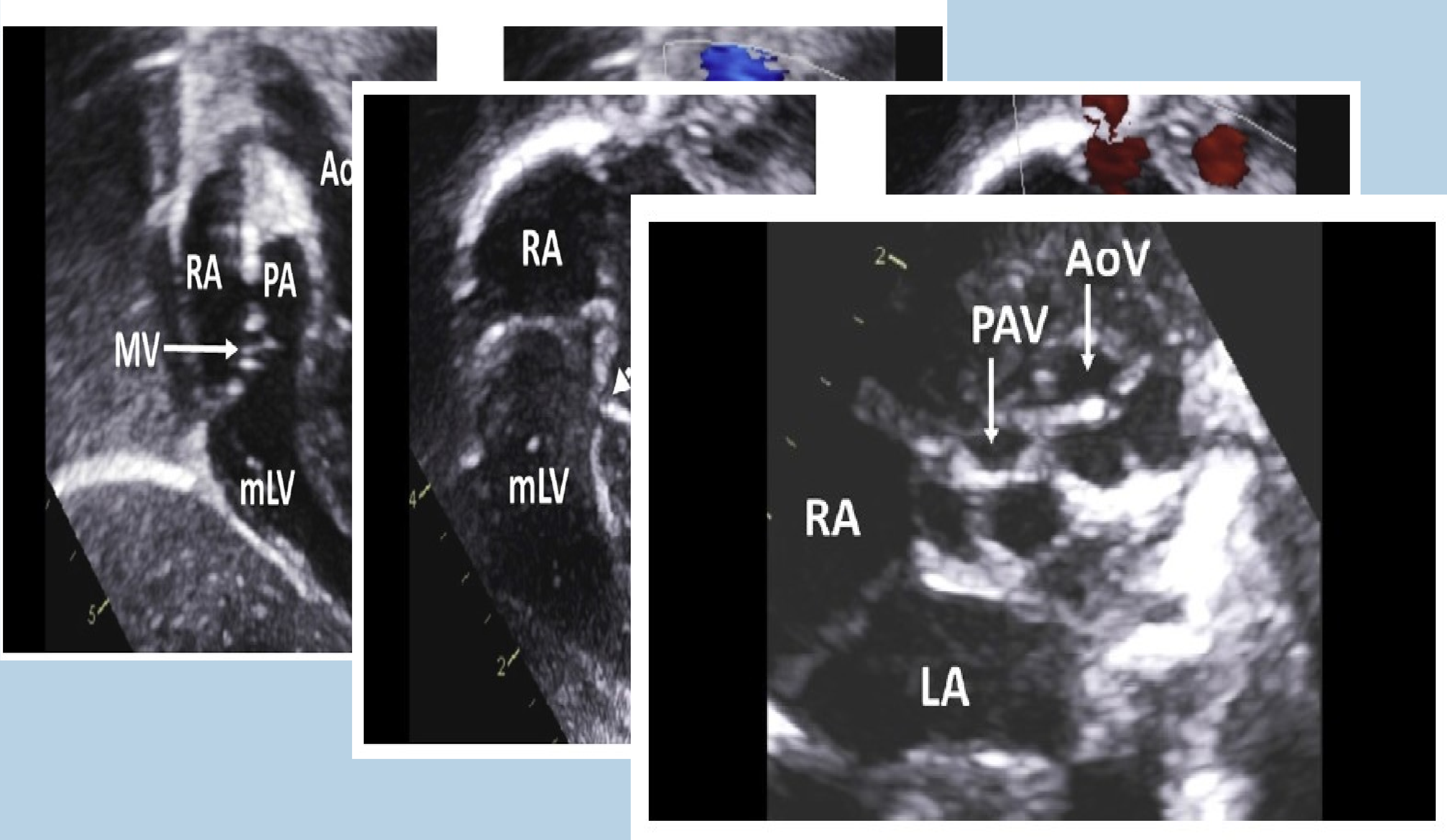
  TR: HF symptoms

  Heart Block: HF symptoms.

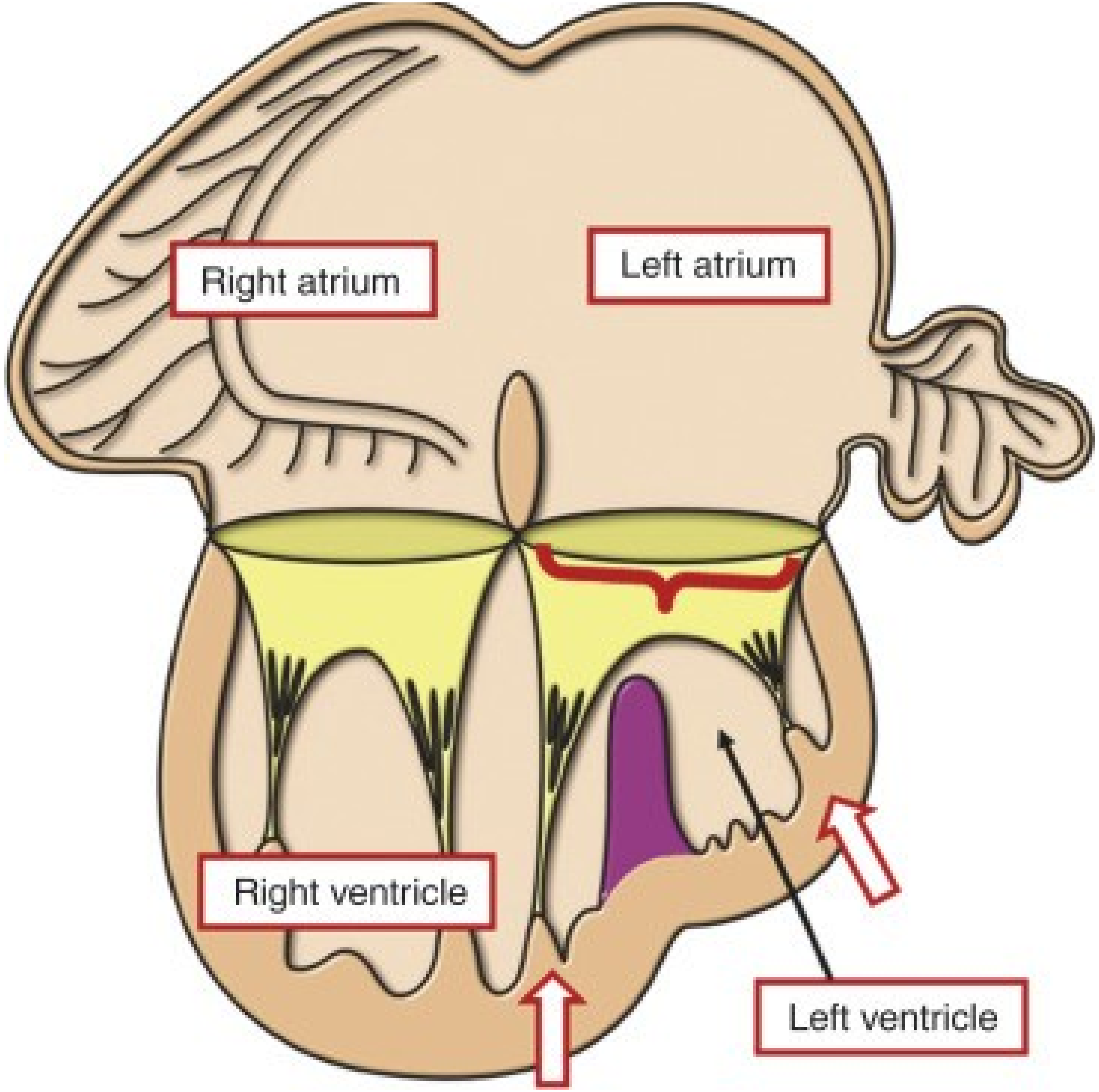
Imaging

- Situs & cardiac position: SI ~10% .. Dextrocardia or Mesocardia ~25%
- Atria and AV connection: discordance
- Ventricles: looping, VSDs, volumes and function, AVV straddling.
- Tricuspid valve: Ebstein-like, incompetence
- Outflow tract obstruction: detection of PS & its level and type (resectable or not).
- Great vessels relationship: Parallel (double-circle in PSAX), aorta is not in fibrous continuity with AVV, PV is wedged between MV and IVS.
- Coronary anomalies.


RA
LA
LV
RV
PA
Ao
RV
LV


RA
PA
MV
mLV
RA
mLV
AoV
PAV
RA
LA

AVV straddling


Right atrium
Left atrium
Right ventricle
Left ventricle

Surgical Options

- Palliative:

  MBT or PDA stent: PS & PA.

  PAB: VSD & RV-TR & bridge to DSO

  May be the definitive therapy.

- Physiological: (systemic RV >> long term RV failure)

  TV repair – VSD closure – PS relief

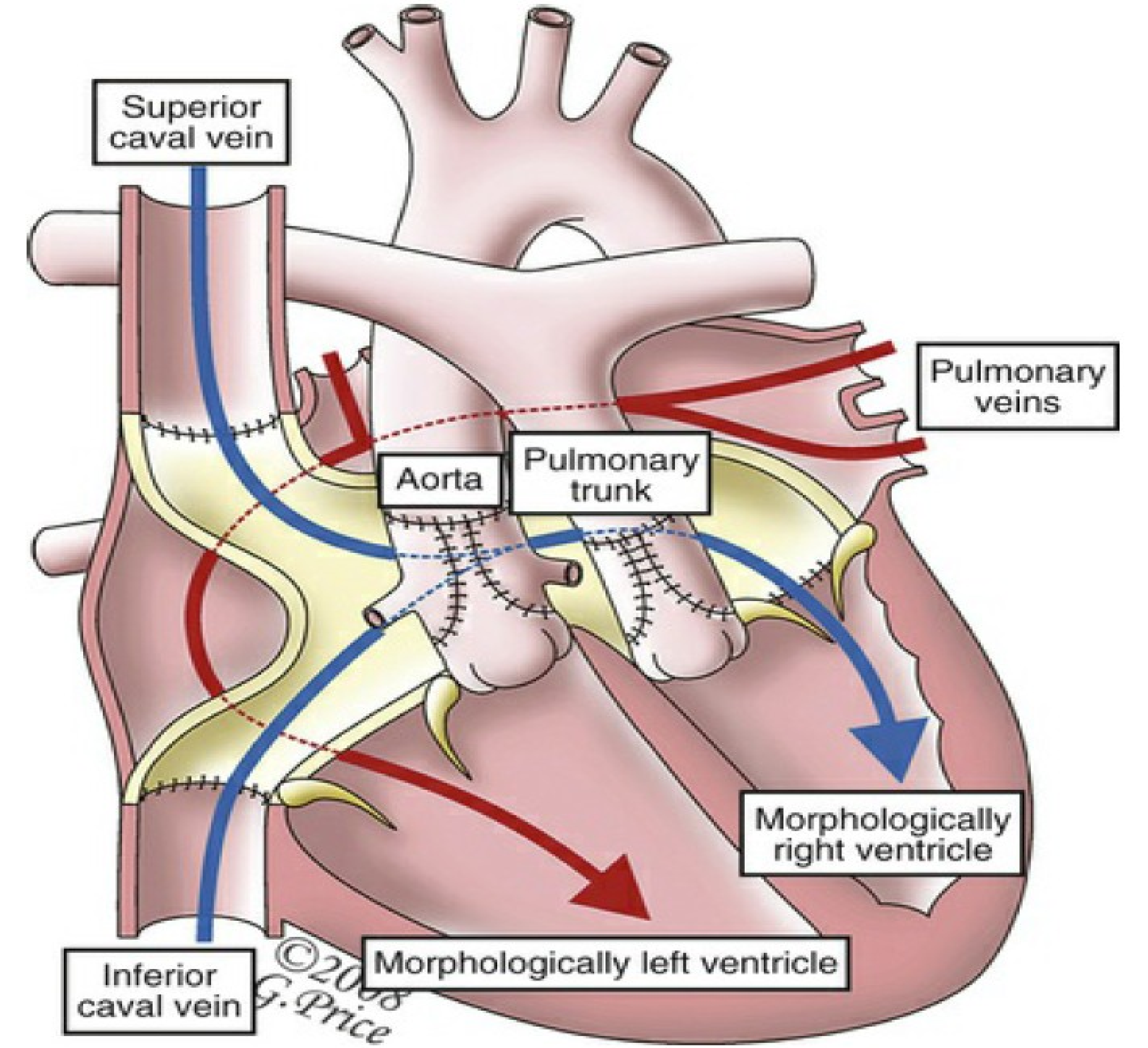
- Anatomical: (systemic LV >> freedom of RV failure)

  DSO – Rastelli – one and half

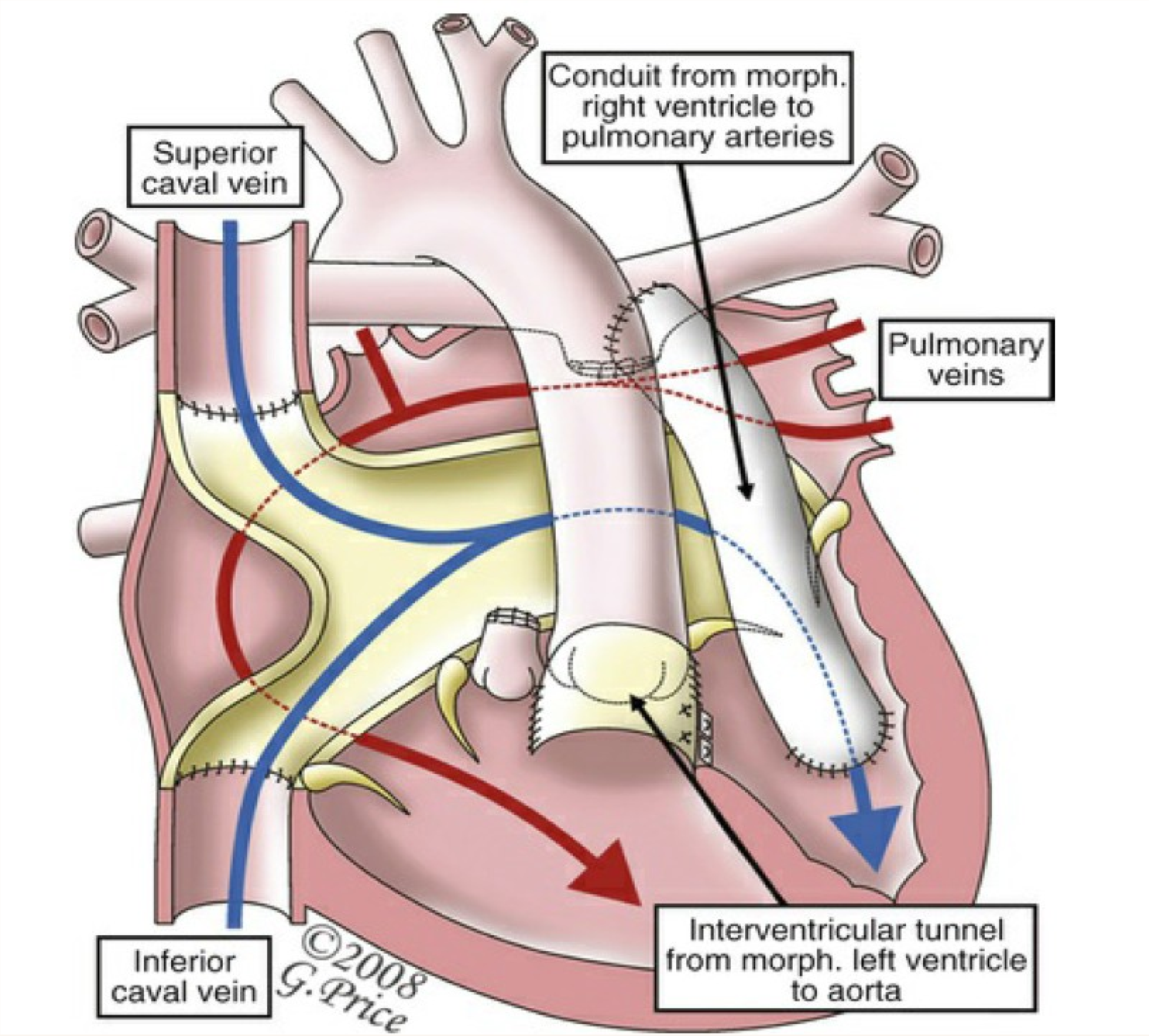
- Single ventricle pathway:

  AVV straddling - hypoplastic ventricle

DSO


Superior caval vein
Pulmonary veins
Aorta
Pulmonary trunk
Morphologically right ventricle
Inferior caval vein
©2008 G. Price
Morphologically left ventricle

Rastelli


Superior caval vein
Conduit from morph. right ventricle to pulmonary arteries
Pulmonary veins
Inferior caval vein
©2008 G. Price
Interventricular tunnel from morph. left ventricle to aorta

Thanks